

## Short Term Prophylaxis

### Minor Manipulations

**If plasma-derived C1 inhibitor (pdC1INH) immediately available:**

- No prophylaxis needed

**If pdC1INH not available:**

Prophylaxis for five days before and two to 5 days post event

- Danazol (avoid during first two trimesters of pregnancy; 2.5-10 mg/kg/day, maximum 600 mg daily)
- Stanozolol 4-6 mg/day

### Major Procedures or Intubation:

**Plasma-derived C1 inhibitor (pdC1INH)**

Give one to six hours before procedure\*  
(optimum dose not yet established – see text)  
Second dose of pdC1INH should be immediately available

**If pdC1INH not available:**

Danazol prophylaxis as per minor and Solvent/detergent treated plasma (SDP; if not available, then fresh frozen/frozen plasma but less safe than SDP) one to six hours before procedure\*  
10 ml/kg; 2-4 units (400-800 ml) for an adult  
**\*as close to procedure as feasible**

## Long Term Prophylaxis

### Plasma-derived C1 Inhibitor

If failing on demand therapy, then continuous pdC1INH prophylaxis twice weekly

### Androgens

Danazol ( $\leq 200$  mg/day)  
Or Stanozolol ( $\leq 2$  mg/day)  
(use lowest effective dose)

### Antifibrinolytic Agents

Less effective than androgens  
Tranexamic Acid (TA)  
20-50 mg/kg/day split bid or tid  
(3-6 g/day maximum)  
(Epsilon aminocaproic acid, EACA if TA not available)